Innovative oxygen therapy saves unborn baby with deadly heart defect

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Linda Luna holds her son, Liam, who was born at Lucile Packard Children's Hospital Stanford with a rare heart defect called congenital Ebstein's anomaly. Credit: Robert Dicks

Prenatal oxygen treatment plus fast and aggressive action after birth helped a San Jose baby born at Lucile Packard Children's Hospital Stanford survive until he could undergo surgical repair of his heart at 11 days old.

Linda Luna was five months pregnant with her first child when she got the bad news: Ultrasound scans showed a deadly defect in her baby boy's heart. He had a 90 percent chance of dying before or just after birth.

But thanks to a groundbreaking treatment at Lucile Packard Children's Hospital Stanford, baby Liam, who went home to San Jose in January, is beating those odds. Now 3 months old, he is thought to be the first baby in the world successfully treated with prenatal maternal hyperoxygenation for his rare heart defect: congenital Ebstein's anomaly.

The problem at diagnosis? Because of severe leaks in two heart valves, blood flowed backward through the right half of Liam's heart. His heart became dangerously enlarged. Too little blood reached his lungs and the rest of his body. Left untreated, the defect would cause irreparable heart and lung damage.

Devising a new approach

"Once you see the type of leakage Liam had, it's usually a progressive process," said Theresa Tacy, MD, associate professor of pediatrics at the School of Medicine and Liam's fetal cardiology specialist. "It just gets worse: The fetus eventually develops heart failure and dies."

To improve Liam's prognosis, Tacy wanted to change his blood flow before birth. In two prior cases of Ebstein's anomaly, she had administered a medication called indomethacin to the pregnant mothers. The drug, an anti-inflammatory similar to ibuprofen, constricted a specific blood vessel near the fetal heart, causing more blood to flow

toward the fetal lungs. It helped both babies to survive until birth, but both died soon after.

So this time, Tacy worked with a team of more than 30 experts from a variety of disciplines at the hospital's Fetal and Pregnancy Health Services to devise a new approach: Expectant mom Luna received 12 hours per day of high-flow oxygen through a face mask, starting when she was 33 weeks pregnant. The idea was to relax Liam's lung blood vessels with the extra oxygen he'd get from his mom. This would make it easier for his heart to pump blood forward into his lungs and, the doctors hoped, help him survive birth and the first few days of life until he was strong enough to have his heart surgically repaired.

More bad news

At the same time they were devising Liam's treatment plan, the team observed a worrying decline in Luna's liver function. Her symptoms could signal a dangerous pregnancy complication known as mirror syndrome.

"We couldn't tell if mom was sick because of the baby's heart problem, which would indicate mirror syndrome, or if she had liver dysfunction for other reasons," said Luna's high-risk obstetrician, Katherine Bianco, MD, associate professor of obstetrics and gynecology. Mirror syndrome can lead to an obstetric emergency that requires immediate delivery of the baby, Bianco added. Because of the risky symptoms, Luna stayed in the hospital's antepartum unit for round-the-clock monitoring.

"Linda was always very positive and willing to do whatever it took, even though we told her so many times 'We don't think this baby is going to make it,'" Bianco said.

"We were trying to offer Liam's parents hope but also remain realistic,"

said David Axelrod, MD, assistant professor of pediatric cardiology. "We knew that even if he made it through pregnancy, his risk of dying during his first few days of life was very high."

On the morning of Nov. 22, about a month before her due date, Luna woke up having contractions. Liam's heart rate was unstable, and the physicians decided he should be delivered by C-section. He had received about three weeks of the prenatal oxygen therapy.

"I was very scared," Luna said. "But the medical team said 'We've been ready, we've been watching him closely and you're ready to have him.'"

Oxygenating the blood

Liam was born that day at 5:47 p.m. The doctors immediately put him on an extracorporeal membrane oxygenation machine to deliver oxygen to his blood. Frank Hanley, MD, professor of cardiothoracic surgery, also surgically closed a blood vessel near the baby's heart to help Liam's blood to flow forward. The team was in such a hurry that Liam wasn't even weighed at birth; saving his life took precedence. Once Liam was stable, Luna's husband, Jose Silva, returned to the recovery room to reassure her that their newborn had cleared an important hurdle.

"Jose told me that Liam's heart was doing it on its own, but he needed help breathing," Luna said. Around midnight, she visited Liam for the first time in the cardiovascular intensive care unit. "I didn't see him until he had his chest open and was on ECMO," she said. "It was very hard, the first time seeing my son, to see him like that."

"After he was born, Liam was severely ill," Axelrod said. Keeping newborns on ECMO is dangerous because they require blood thinners that put them at risk of bleeding and stroke. And the ECMO machinery can expose them to air bubbles and tiny blood clots that would further

increase stroke risk. Yet without ECMO, Liam's body wouldn't get enough oxygen. Balancing the risks was "like trying to catch a falling dagger," Axelrod said.

Finally, by Dec. 3, Liam's lungs were working well enough that the physicians thought he could probably breathe on his own. In surgery that day, Hanley extubated Liam and fully repaired his heart, fixing the malfunctioning valves and reducing the size of the enlarged right atrium.

"It was a huge surgery for a tiny baby fighting for his life," said Luna. "The seven-hour wait during surgery was the longest wait of my life, but when they finally wheeled him out, he was a different baby. We were so thankful."

Liam is now doing well. He will be monitored by a cardiologist as he grows, but the physicians expect he will otherwise be an ordinary kid. "He's got a great outlook, which is a really satisfying thing," Axelrod said. "Our team did a great job."

Added Tacy, "This is just one case, but it's so exciting to move forward and feel a glimmer of what we think is the right path for treating other babies with this devastating heart defect."

Provided by Stanford University Medical Center